Treatment of Intracranial Venous Occlusive Disease with Sigmoid Sinus Angioplasty and Stent Placement in a Case of Infantile Multifocal Dural Arteriovenous Shunts

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Summary

The infantile dural arteriovenous shunts are multifocal involving different dural sinuses and progress to an occlusive venopathy with sigmoid sinus and/or jugular bulb stenosis and subsequent occlusion. We report a successful angioplasty and stent placement of a sigmoid sinus - jugular bulb stenosis due to venous occlusive disease in a patient with infantile dural arteriovenous shunts.

A five-year-old patient presented with status epilepticus due to severe venous congestive encephalopathy. The angiogram revealed multifocal dural arteriovenous shunts, occlusion of the right sigmoid sinus, absence of cavernous sinuses and significant stenosis of the left sigmoid sinus - jugular bulb.

By transvenous approach, percutaneous transluminal balloon angioplasty and stent placement of the stenosed left sigmoid sinus - jugular bulb segment was performed. This resulted in a significant decrease of the venous pressure gradient across the stenosis and allowed a dramatic clinical recovery.

Dural sinus angioplasty and stent placement appears to be a safe and effective procedure and should be considered in the treatment of the venous occlusive disease associated with infantile dural arteriovenous shunts.

Introduction

Intracranial dural arteriovenous shunts are a group of different nosologic entities sharing a similar angioarchitecture. They have been classified by Lasjaunias et Al as adult-type dural arteriovenous shunts, infantile dural arteriovenous shunts and dural malformations with arteriovenous shunts [1]. In the paediatric population, intracranial dural arteriovenous shunts are rare, accounting for less than ten percent of all types of arteriovenous shunts [1].

Infantile dural arteriovenous shunts are high flow and multifocal involving different dural sinuses [1,2,3]. The multiplicity of shunts interferes with the venous drainage of the normal brain leading to venous congestive encephalopathy [1]. The natural course of the disease includes a progressive occlusive venopathy with sigmoid sinus and/or jugular bulb stenosis and subsequent occlusion [4]. This exacerbates the venous

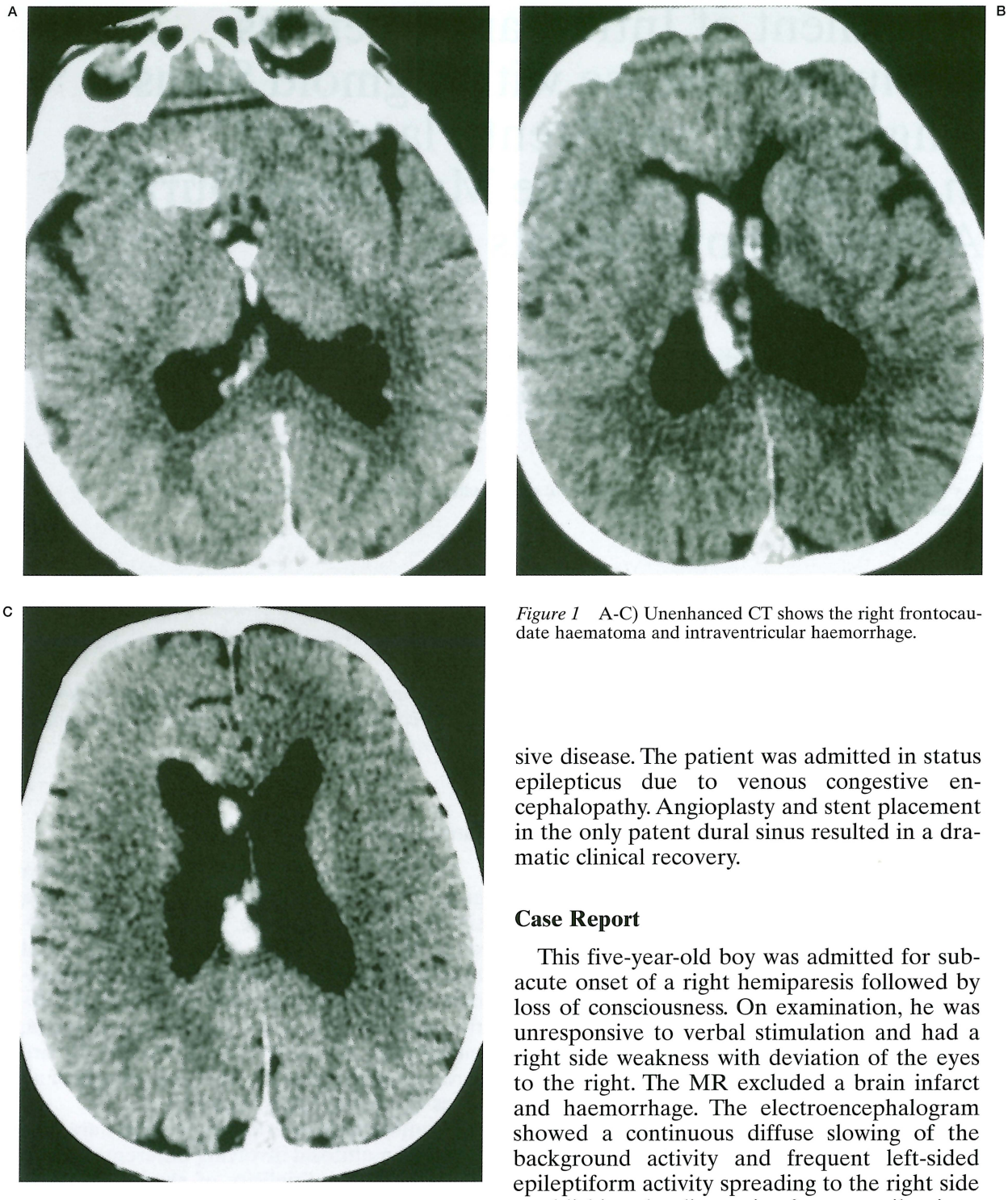

Figure 1 A-C) Unenhanced CT shows the right frontocaudate haematoma and intraventricular haemorrhage.

congestion in the brain and may change the venous drainage pattern of the dural shunts, with the development of cortical venous reflux[4].

The authors report a case of infantile dural arteriovenous shunts with severe venous occlusive disease. The patient was admitted in status epilepticus due to venous congestive encephalopathy. Angioplasty and stent placement in the only patent dural sinus resulted in a dramatic clinical recovery.

Case Report

This five-year-old boy was admitted for subacute onset of a right hemiparesis followed by loss of consciousness. On examination, he was unresponsive to verbal stimulation and had a right side weakness with deviation of the eyes to the right. The MR excluded a brain infarct and haemorrhage. The electroencephalogram showed a continuous diffuse slowing of the background activity and frequent left-sided epileptiform activity spreading to the right side establishing the diagnosis of status epilepticus. This was refractory to combined anti-seizure therapy and required intubation with the institution of phenytoin, phenobarbarbital and midazolam.

This patient had presented early at the age of

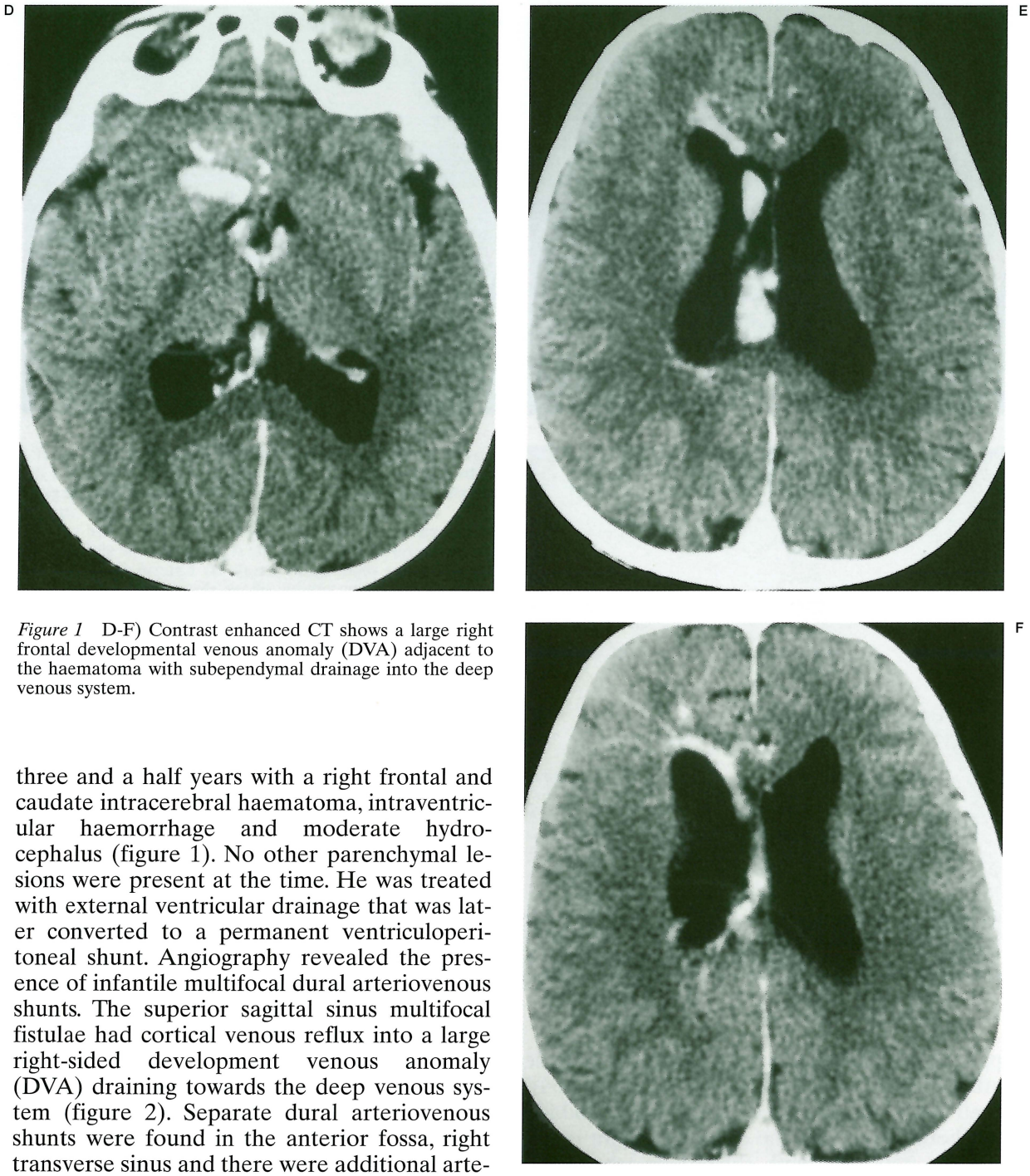

Figure 1 D-F) Contrast enhanced CT shows a large right frontal developmental venous anomaly (DVA) adjacent to the haematoma with subependymal drainage into the deep venous system.

three and a half years with a right frontal and caudate intracerebral haematoma, intraventricular haemorrhage and moderate hydrocephalus (figure 1). No other parenchymal lesions were present at the time. He was treated with external ventricular drainage that was later converted to a permanent ventriculoperitoneal shunt. Angiography revealed the presence of infantile multifocal dural arteriovenous shunts. The superior sagittal sinus multifocal fistulae had cortical venous reflux into a large right-sided development venous anomaly (DVA) draining towards the deep venous system (figure 2). Separate dural arteriovenous shunts were found in the anterior fossa, right transverse sinus and there were additional arteriovenous shunts in the falx cerebri (figure 2, 3). The right sigmoid sinus was occluded and there was a diffuse venous congestion of the brain (figure 2, 3). The patient at that time recovered without neurological deficit.

Over the next twenty-three months, the patient had five embolisations of the multiple fistulae. The transarterial embolisations were done using polyvinyl alcohol particles (PVA) and N-butyl- cyanoacrylate (NBCA). The superior sagittal sinus fistula was also treated with coil packing of the involved sinus. These embolisations resulted in some reduction of the arteriovenous shunts.

During this twenty-three month period, the

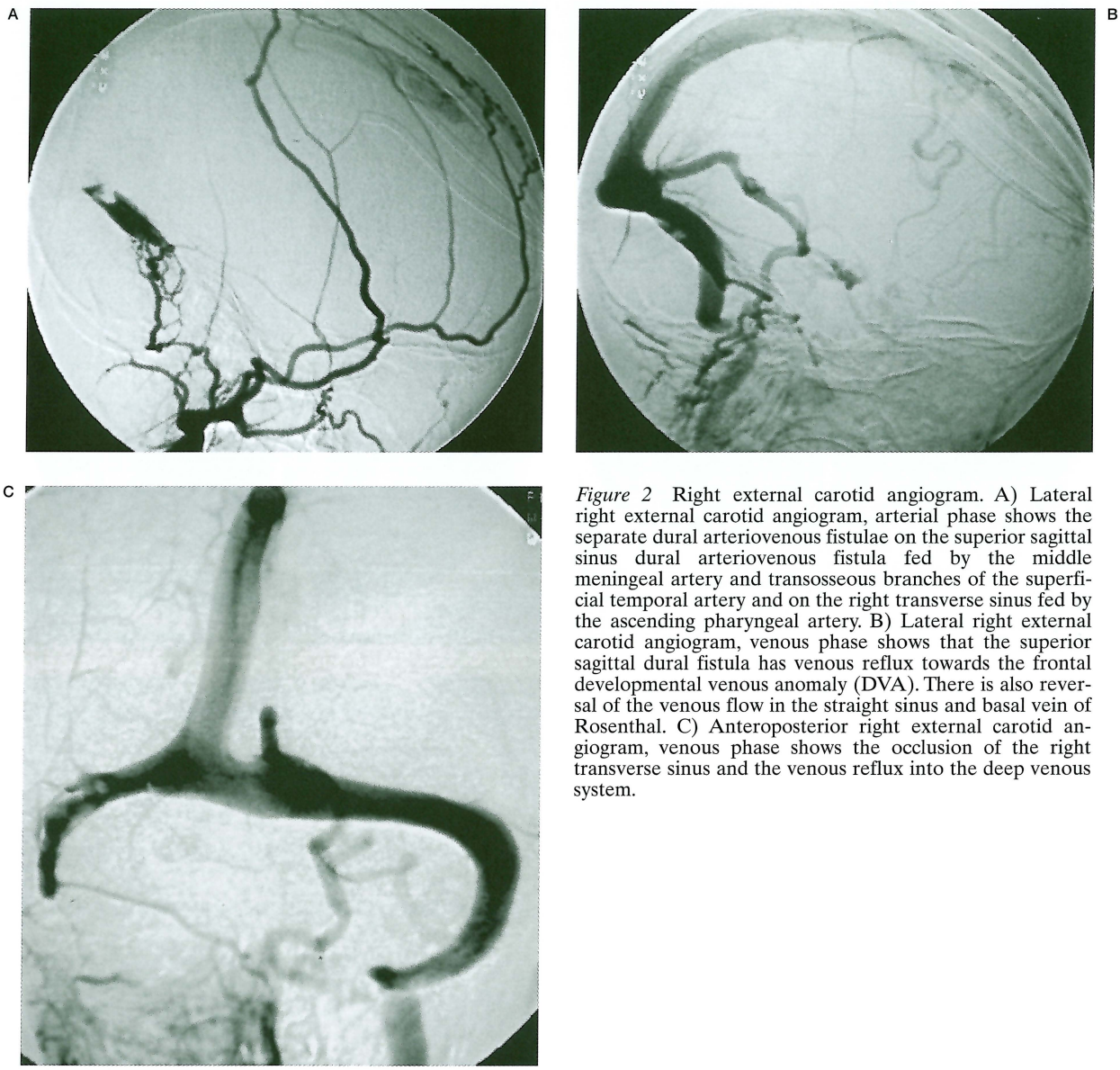

Figure 2 Right external carotid angiogram. A) Lateral right external carotid angiogram, arterial phase shows the separate dural arteriovenous fistulae on the superior sagittal sinus dural arteriovenous fistula fed by the middle meningeal artery and transosseous branches of the superficial temporal artery and on the right transverse sinus fed by the ascending pharyngeal artery. B) Lateral right external carotid angiogram, venous phase shows that the superior sagittal dural fistula has venous reflux towards the frontal developmental venous anomaly (DVA). There is also reversal of the venous flow in the straight sinus and basal vein of Rosenthal. C) Anteroposterior right external carotid angiogram, venous phase shows the occlusion of the right transverse sinus and the venous reflux into the deep venous system.

patient had five episodes of transient ischaemic attacks (TIA) each lasting less than one hour. Four were characterized by transitory sudden left hemiparesis and the other by a right hemiparesis associated with slurred speech. In the acute phase of one of these events an electroencephalogram was performed, ruling out the possibility of an ictal nature. The patient was asymptomatic between these events with normal developmental milestones. CT at four and a half years of age showed the development of brain calcifications, mainly in the frontal lobes.

During the current admission, the cerebral angiogram revealed a significant left sigmoid sinus and jugular bulb narrowing which had progressed from the original angiogram (figure 4). The right transverse sinus fistula had recruited new arterial feeders and there was now venous reflux towards the perimedullary venous system. The cavernous sinuses had not developed. The patient was started on IV heparin (20 U / Kg / h) that afterwards was replaced by subcutaneous low molecular heparin (Enoxaparin 0.75 mg / Kg / bid). Two embolisations were done to treat the right transverse sinus fis-

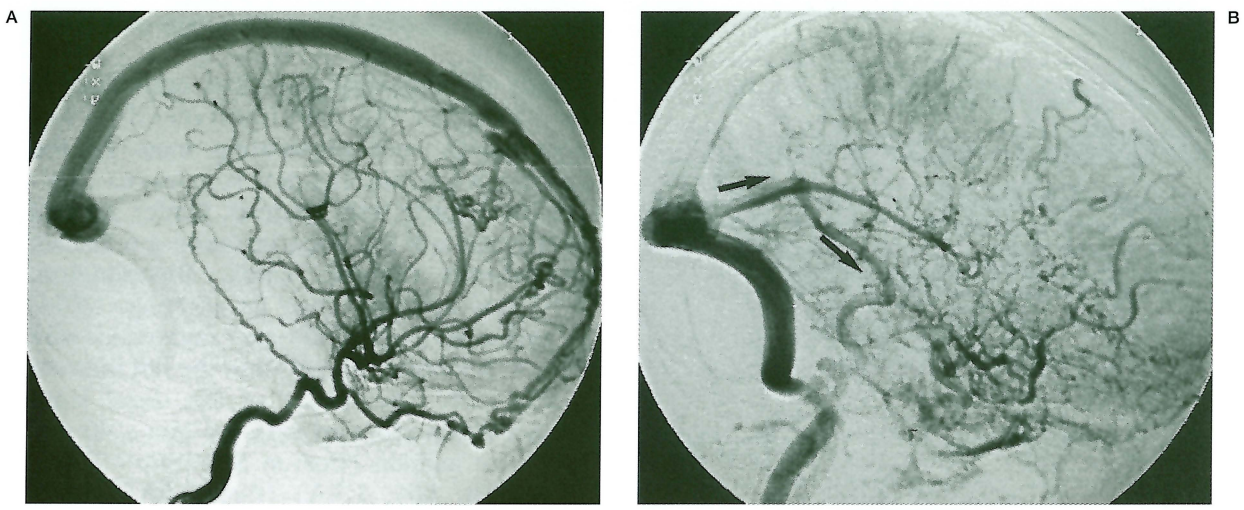

Figure 3 Right internal carotid angiogram. A) Lateral right internal carotid angiogram, arterial phase shows an anterior fossa dural AV shunt fed by dural branches of the ophthalmic artery and two separate arteriovenous shunts in the falx cerebri fed by anterior cerebral artery branches. An enlarged Bernasconi-Cassinari artery is supplying the superior sagittal sinus fistula. B) Lateral right internal carotid angiogram, venous phase shows the venous congestion of the brain with dilated and tortuous frontal and sylvian veins and reversal of the flow in the straight sinus, vein of Galen and vein of Rosenthal (arrows).

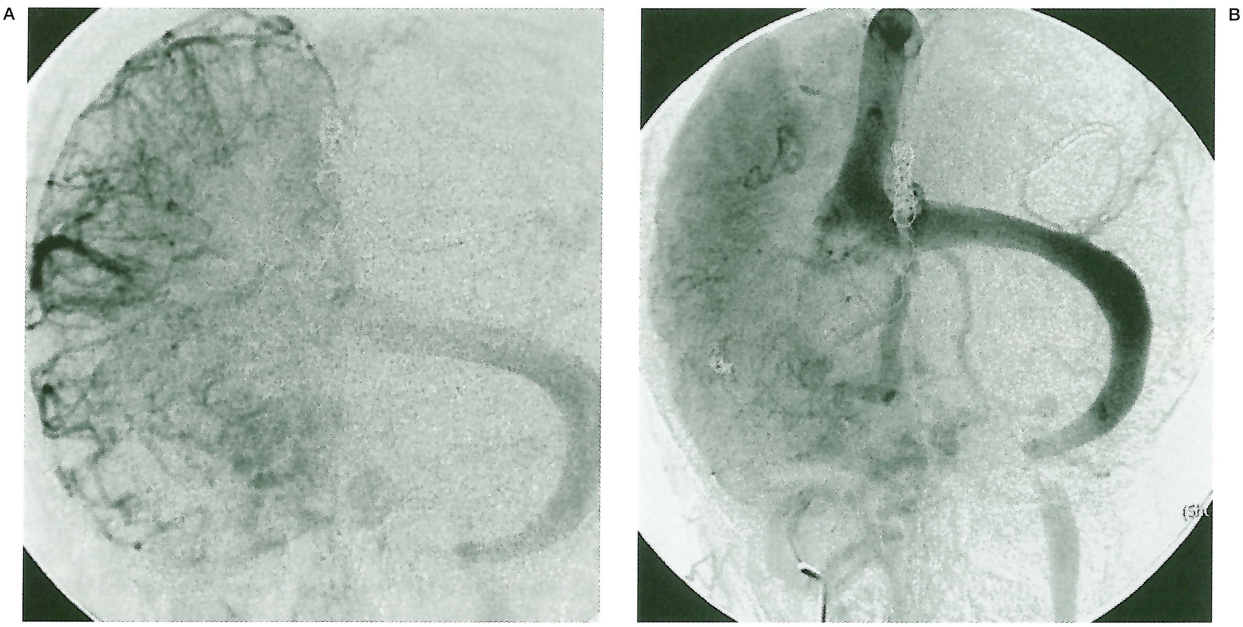

Figure 4 Left sigmoid sinus - jugular bulb narrowing. A) Angiogram performed nineteen months after the initial presentation. Anteroposterior right internal carotid angiogram, venous phase shows a slight increase in the left sigmoid sinus - jugular bulb narrowing comparing to the previous angiograms (see figures 2 and 3). B) Angiogram performed during the current admission, before the angioplasty and stent placement (twenty-three months after the initial presentation). Anteroposterior right internal carotid angiogram, venous phase shows a strikingly increase in the left sigmoid - jugular bulb stenosis.

tula. This included arterial embolisations, and transvenously, disconnection of the cortical venous reflux with coils. A pressure gradient of 34 mmHg was recorded across the left sigmoid sinus / jugular bulb stenosis with a venous blood pressure of 47 mmHg proximal and 13 mmHg distal to the stenosis. After these treatments, there were still residual dural arteriovenous shunts involving the right transverse sinus and falx cerebri. After these embolisations the pa-

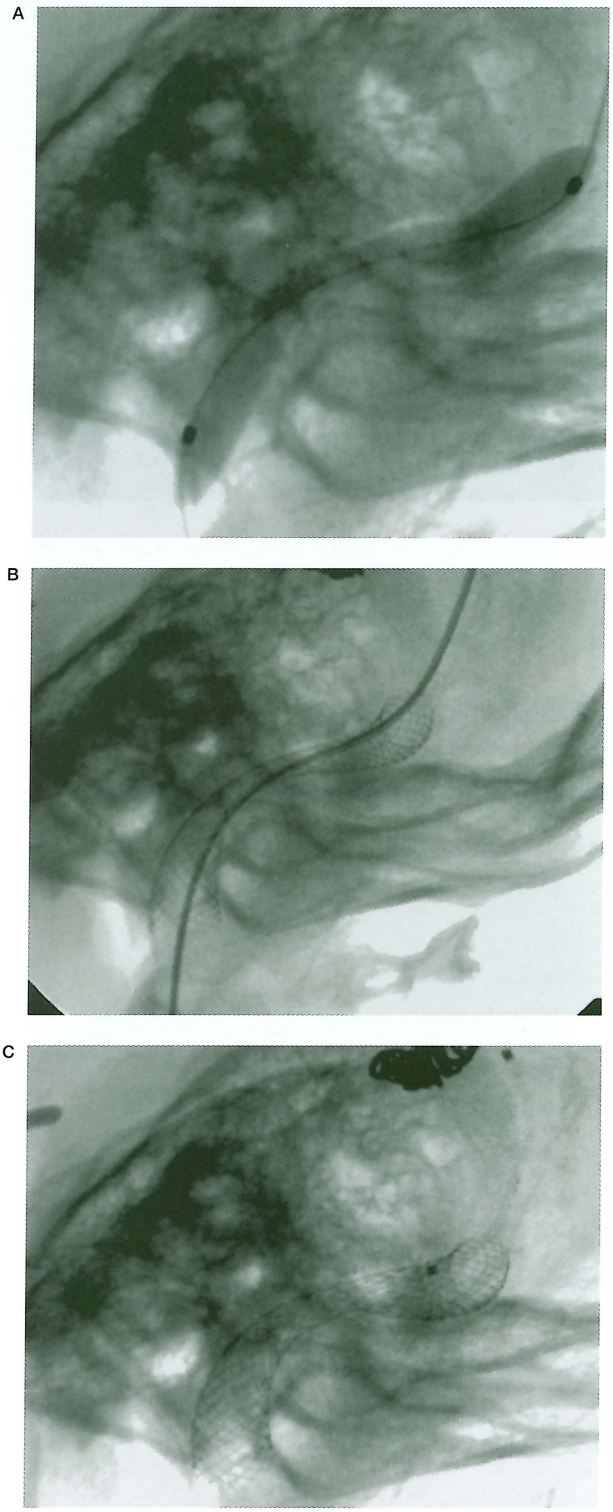

Figure 5 Lateral radiographs during angioplasty and stent placement. A) Shows the inflated PTA balloon within the left sigmoid sinus and internal jugular vein. B) Shows the stent within the sigmoid sinus and internal jugular vein before post-dilation of the stent. C) Shows the final result after stent placement and post-dilation.

tient was extubated and made a slow neurologic recovery.

Since the recovery was slow, embolisation was done again. The right transverse sinus was packed with coils eliminating most of the shunting at this site. The next step was to eliminate the jugular bulb stenosis. Under general anaesthesia and full heparinization a 6 French long sheath (Shuttle, Cook, Bloomington, USA) was advanced over an exchange 0.014-inc exchange-length (300 cm) microguidewire (Boston Scientific / Target) into the left internal jugular vein. Angioplasty of the dural sinus stenosis was performed with a 4 mm x 40 mm Symmetry balloon (Boston Scientific / Target Watertown, MA, USA) (figure 5A). Afterwards, an 8 mm x 20 mm Wallstent Reduced Profile (Boston Scientific / Medi-tech, Minneapolis, MN, USA) was advanced over the Amplatz exchange-length (300 cm) 0.035-inch wire (Cook, Bloomington, USA) and placed over the stenosed segment (figure 5B). The stent was post-dilated with a 5 mm x 20 mm Ultra-thin Diamond balloon (Boston Scientific / Target / Medi-tech, Watertown, MA, USA) (figure 5C). Finally, a 5 French guiding catheter was introduced in the transverse sinus and the venogram confirmed the successful reopening of the stenosis (figure 6). The venous pressure gradient was reduced to 14 mmHg.

The patient was started on acetylsalicylic acid (AAS) two days before the procedure and on clopidogrel (18.75 mg / day) after the procedure and during the following five days. Low molecular heparin was maintained during the admission and AAS was continued indefinitely. The patient became more alert with a progressive improvement in motor function and cognitive ability. At three month follow-up, the patient had recovered from his neurological deficits and had resumed his usual school activity. The venous ultrasonography doppler showed the patency of the sigmoid sinus - internal jugular vein junction with presence of an antegrade in the internal jugular vein at the level of the skull base.

Discussion

The prognosis of infantile dural arteriovenous shunts depends on the multifocality of the dural shunts and the progression of the venous occlusive disease. Multifocal dural arteriove-

nous shunts may lead to a venous congestive encephalopathy that will be exacerbated by the progression of the dural venous occlusive disease. Rerouting of the venous drainage towards uninvolved venous pathways, like the cavernous sinus, can occur. This may delay the deleterious effects of the venous congestion but will create additional symptoms related to venous congestion in the re-routing venous pathways[4].

Treatment of infantile dural arteriovenous shunts is difficult and complex. The aim is to occlude the dural shunts and maintain the patency of the venous outlets of the brain.

Transarterial embolisation with NBCA is often the initial treatment. It is usually partial as the multifocal nature of the disease leads to the early development of new arteriovenous shunts near the previously embolised areas. The transvenous approach with endovascular dural sinus sacrifice is often not possible, nor recommended, due to the involvement of multiple sinuses by this disease[4].

Our case of infantile dural arteriovenous shunts represented a major challenge. The patient presented with an intracranial haemorrhage, which is expected in the final stages of the disease. The venous drainage of the multifocal superior sagittal sinus dural fistulae was refluxing towards a large frontal DVA. Since the DVA may be a weak anatomical variant, we suspect it was, in part, responsible for the initial haemorrhage.

Transient ischaemic attacks (TIA) are not common in the setting of venous congestion. We postulate that the TIAs resulted from sudden and transitory increases in the intracranial venous pressure. This leads to a decrease of the arterial flow to the brain, which is subjected to the cortical venous reflux. An elevated central venous pressure can create a significant decrease in the cerebral blood flow, especially if a compensatory increase in the mean arterial pressure does not occur[5,6].

A significant venous congestion of the brain was present from the time of diagnosis in our patient. The venous drainage of the superior sagittal sinus dural shunts was also refluxing towards the deep venous system. Although, the patient remained asymptomatic for months after presentation, the follow-up CT demonstrated the development of parenchymal calcifications related to the progressive venous congestive encephalopathy.

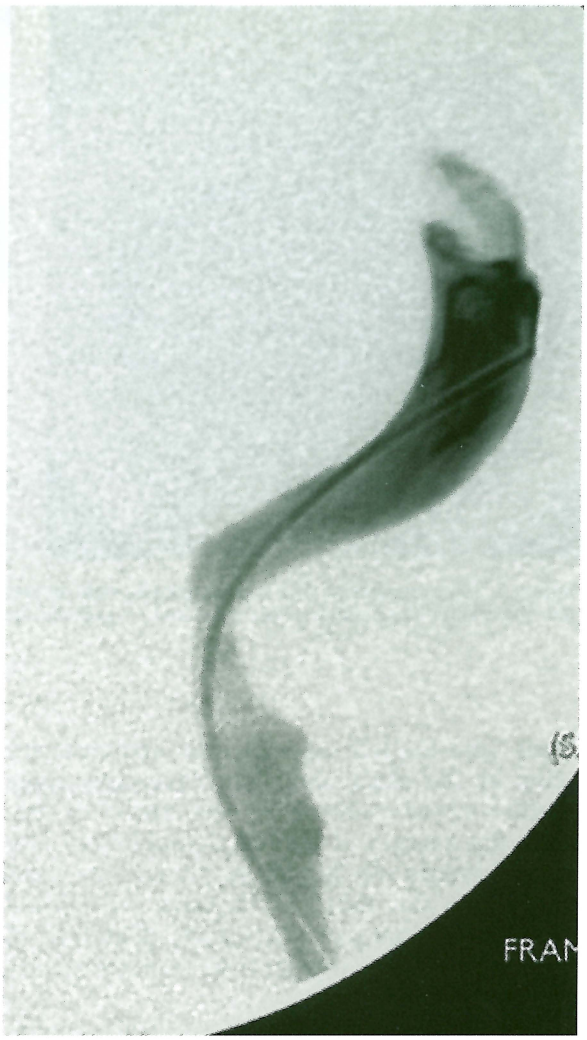

Figure 6 Anteroposterior left sigmoid sinus venogram after angioplasty and stent placement shows significant dilatation of the sigmoid sinus - jugular vein stenosis after treatment with angioplasty and stent placement.

Staged transarterial embolisations helped to reduce the dural shunts and relieve the venous congestion. However, the natural history of the disease leads to early recurrence of the dural shunts and the benefit was transient. The transvenous approaches, with occlusion of segments of the dural sinus not used by the normal brain venous drainage, allowed additional relief of the venous congestion and disconnection of the pial cortical venous reflux. This reduced the haemorrhagic risk.

A dramatic clinical deterioration occurred when the left sigmoid sinus became progressively occluded. The only venous outlets of the

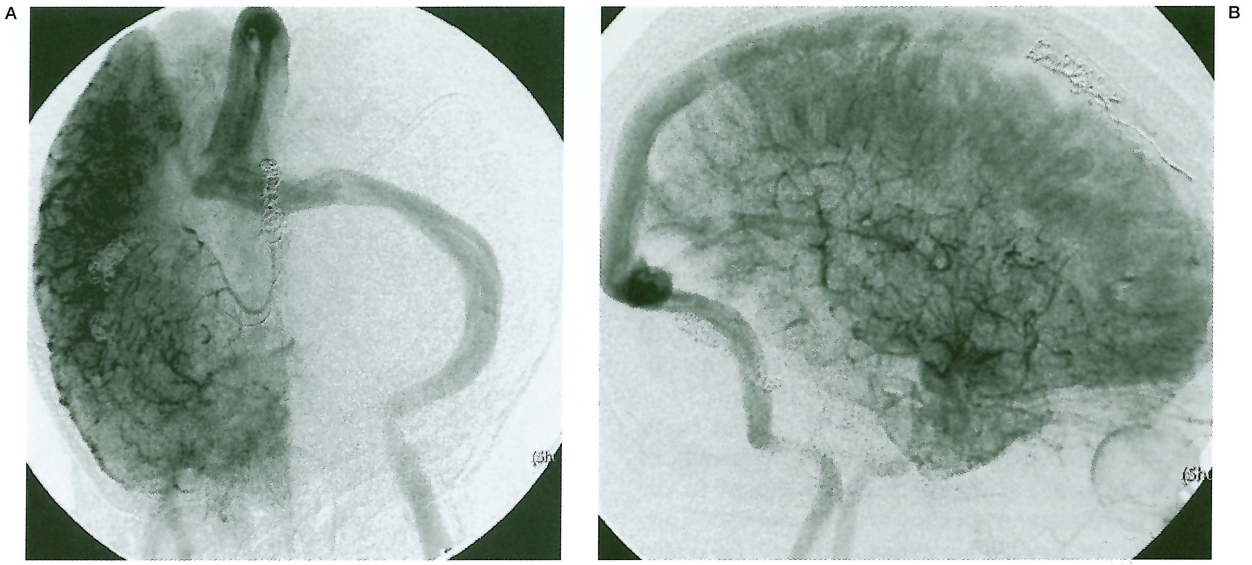

Figure 7 Anteroposterior (A) and lateral (B) right internal carotid angiogram, venous phase, after angioplasty and stent placement show the patency of the stent and a significant increase in the diameter of the sigmoid sinus - jugular vein lumen.

brain were the left sigmoid sinus and emissary veins in the skull base. The cavernous sinus capture never occurred. It was crucial to preserve the patency of the left sigmoid sinus. The use of heparin was helpful to prevent the sinus thrombosis but failed to improve the clinical status of the patient. Therefore, dilatation of the sigmoid sinus / jugular bulb stenosis was required.

The majority of the data regarding angioplasty and stent placement in the venous system come from the treatment of extracranial peripheral and/or central venous stenosis and thrombosis [7-13]. Central venous stenoses are particularly common in haemodialysed patients. This is partially due the high flow state associated with AV fistula leading to endothelial and fibromuscular hyperplasia in areas of flow turbulence and shear stress [8,10,11]. Central venous stenosis responds to angioplasty but there is a high incidence of early recurrence [7-12]. This is probably due to venous recoil, which is more common in elastic lesions [10]. The use of stent prevents early recurrence and prolongs the interval of time without the need for re-interventions [8,9,10,12]. Some of the technical complications of venous stent placement include venous perforation, stent migration or misplacement and early or delayed stent shortening leaving an uncovered stenosis [7,8,12]. The causes for restenosis after stenting are thrombosis, intimal hyperplasia and venous stenosis adjacent to the ends of the stent [9]. In the larger series of haemodialysed patients with venous stenosis treated with stents, the reported primary patency rate is 56% and 28% at one and two years, respectively; but the cumulative primary patency rate was 81% at four years [8]. In the treatment superior vena cave stenosis due to benign disease, the primary and cumulative primary patency rates were 77% and 85% respectively, at a mean follow-up of 17 months [13]. Recently, primary and cumulative primary patency rate of 80% and 87%, at a mean follow-up of 19 months, were reported in the treatment of inferior vena cave stenosis [7].

The experience of dural sinus angioplasty and stent placement is anecdotal [14,15,16]. In 1994, Marks et Al reported the first two cases of angioplasty and stent placement in a dural sinus. The patients had symptomatic dural sinus stenosis not related to arteriovenous shunts. The angioplasty and stent placement resulted in a dilatation of the dural sinus and there was a decrease in the blood pressure gradient through the stenosis [14]. Malek et Al reported a case of spontaneous dural pan-sinus thrombosis refractory to anticoagulant therapy. Subsequently, the 13-year-old patient developed multiple acquired (adult type) dural arteriovenous

fistulae (AVF). Angioplasty and stent placement allowed the reopening of a dural sinus and decreased the venous pressure gradient [15].

Recently, Murphy et Al reported a case of left transverse sinus dural AVF associated with occlusion of the left transverse and sigmoid sinuses. Angioplasty and multiple stent placement, from the transverse sinus to the internal jugular vein, reopened the dural sinuses and produced occlusion of the dural AVF [16]. There is one case report of bilateral traumatic jugular vein thrombosis refractory to thrombolysis successfully treated with stent placement [17]. Intracranial dural sinus angioplasty alone was associated with early recurrent stenosis or occlusion in the three aforementioned case-reports [14,15,16]. The long-term follow-up of the dural sinus treated with stent placement is not known. In the three case-reports mentioned, the clinical or radiological follow-up ranged from 2 to 12 months and in all cases the stents were still functional [14,15,16].

In conclusion, dural sinus angioplasty and stent placement is a safe and effective procedure to treat dural sinus stenosis. This option should be considered in the treatment of venous occlusive disease associated with infantile dural arteriovenous shunts. Angioplasty alone is generally associated with early restenosis or occlusion and therefore stent placement is necessary. Longer follow-up is needed to evaluate the long-term stent patency and efficacy of this treatment.

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EDITORIAL COMMENT

The authors reported the successful insertion of a stent into the marked stenotic segment of the dural sinus as a final option in a five-year-old boy who is resistant to arterial and venous embolization of the fistular tracts. To the best of our knowledge, he is the youngest patient among those reported to receive stenting in the intracranial venous pathway.

This report adds support to the strong evidence that relief of high venous pressure through stenting will become a valuable therapeutic option when there is an uncontrollable fistula leading to venous hypertension due to compromised drainage in the dural sinus.

The timing of the stenting will be the critical issue because the guide-wire or balloon should pass the stenotic or occluded segment of the dural sinus. Once it is occluded, the intimal thickening and organizing thrombus will be replaced by fibrotic thickening of sinus intima or proliferation of elastic laminar of the sinus through which the passage of stenting devices can be very difficult, although the pathogenesis of sinus occlusion and the reason for development of a dural arteriovenous fistula, are still uncertain[1,2].

How the stent will adjust to the foramen in a growing skull will be another concern in this child. A self-expanding stent may be optimal from that point of view, although only long-term follow-up will provide proof.

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